

CASE STUDY

RARE CASE OF AN ISOLATED ENTERIC DUPLICATION CYST IN A TEENAGER

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ABSTRACT: Intestinal duplication cysts are rare congenital anomalies that are usually detected during infancy or early childhood. Though they can occur anywhere in the gastrointestinal tract, small intestine is the commonest site of occurrence. Majority of the enteric cysts have a communication with the adjacent bowel. Completely isolated enteric duplication cysts are a rare variety of duplication cysts having no communication with any part of the adjacent bowel segment and an independent blood supply. These cysts often present with non-specific symptoms like abdominal pain and vomiting thereby, highlighting the importance of ultrasound as an aid to precise and timely diagnosis. Histopathological examination is confirmatory, with excision of the cyst being the preferred treatment. We present a rare case of an isolated intestinal duplication cyst in a teenage boy which remained undetected in childhood.

KEYWORDS: Duplication cysts, abdominal pain, enteric cyst, histopathology, resection

INTRODUCTION:

Duplication of the alimentary tract is a rare congenital anomaly. It can be found anywhere from the mouth to the anus and can be symptomatic or at times be discovered incidentally. W. E. Ladd first introduced the term duplication in 1934. [1] In the gastrointestinal tract the most common sites of duplications are the ileum, esophagus, and colon. Although majority of the duplications are benign, the presence of

ectopic mucosal rests and their potential for getting converted to malignancy remain a concern. [2] We describe a case of enteric duplication of the cystic type in a teenager which was suspected clinically to be a mesenteric cyst.

CASE REPORT:

A thirteen-year-old boy was referred to the department of pediatric surgery of our hospital with the complaints of recurrent abdominal pain

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and distension. The boy had no history of fever, vomiting or altered bowel habits. All the routine blood parameters were normal. On clinical examination, a cystic mass was palpable in the right mid abdomen. Ultrasonography of the abdomen revealed a characteristic double layered wall with an inner hyper echoic mucosa and an outer hypo echoic rim of smooth muscle layer. A well-defined, uniloculated, non-communicating cystic lesion in relation to ileum measuring 3x2cms was noted. With a clinical diagnosis of mesenteric cyst, surgical excision of the cyst was done as the boy was symptomatic with severe pain. The resected specimen was sent for histopathological examination.

The gross examination revealed a short segment of ileum measuring 3.5 cms in length with an attached cystic mass measuring 3x2x1.5 cms on the mesenteric border. The cystic structure was seen abutting the intestinal segment. Cut section showed a uniloculated cyst, which was non-communicating with the intestinal lumen. The cyst cavity was filled with thick mucoid material (**Figure 1A and 1B**).



Figure 1A and 1B: Gross specimen of the excised enteric duplication cyst attached to the segment of ileum.

Microscopic examination of the cyst wall showed an attenuated lining of ileal mucosa resting on a muscularis layer shared by the adjacent normal appearing intestinal segment (**Figure 2A**). The cyst lining was intact all over and non-communicating with the intestinal lumen. The attached ileal segment was unremarkable (**Figure 2B**). A diagnosis of enteric duplication cyst was conferred upon. The boy

had an uneventful postoperative stay and was discharged after five days of hospital stay.

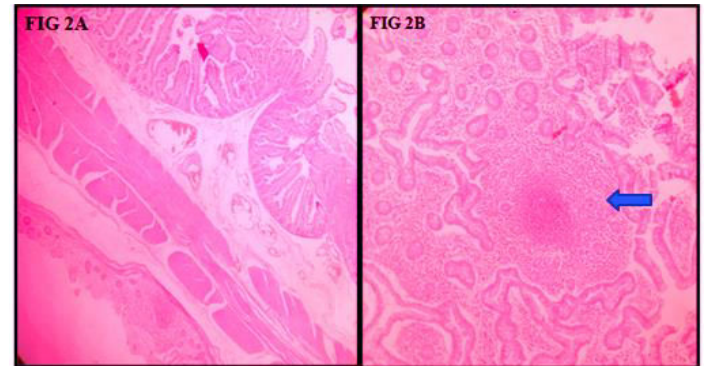


Figure 2A: Histology of the enteric duplication cyst showing two mucosal layers sharing a common submucosal layer with muscular layer. (H & E, 10X). 2B: Histology of the ileal mucosa showing lymphoid aggregate (blue arrow). (H & E, 40X).

DISCUSSION:

Duplications are congenital malformations postulated to arise from disturbances during embryologic development. They are seen in 1:4,500 births, and found in 0.2% of all children, with a slight male predominance.^[3] Intestinal duplications are most commonly detected in infancy and early childhood when symptoms such as abdominal pain, obstruction, or vomiting develop.^[4] Some duplication cysts may remain asymptomatic until adulthood.^[5] The patient in our case was a 13-year-old boy who presented with abdominal pain and distention. He had similar complaints in the past, however coming from a rural background no speciality consultation was done for the same till the patient came to our hospital. Although the boy gave history of similar complaints in the past, coming from a rural background no speciality consultation was done for the same till the patient came to our hospital.

Multiple theories have been proposed for embryological occurrence of duplication cysts in different locations which includes persistence of fetal gut diverticula, defect in solid stage of recanalization of primitive gut and partial twinning of the primitive

gut.^[6] Development of a split notochord that allows connection between the yolk sac endoderm and the ectoderm was proposed as the primary defect by Bentley and Smith, who further added that subsequent duplication of the gut resulted from eventration of the yolk sac between the halves of the vertebra.^[1]

The commonest sites of duplications in the gastrointestinal tract are Jejunum and ileum (40 – 70%), colon (17 %), esophagus (13%), stomach (8%) and duodenum (2-12%) with majority of them being located on the mesenteric side.^[7] Enteric duplications can further be of cystic, tubular or mixed type.^[8] Ours was an enteric duplication of the cystic type, also called an enteric duplication cyst.

Although they are attached to the intestinal wall and share a common blood supply, majority of the duplication cysts rarely communicate with the intestinal lumen. These cysts derive their blood supply from the same mesenteric vessel that supplies the adjacent bowel. The significance of this lies in the fact that a shared blood supply with the neighboring bowel will necessitate a bowel resection as a part of the surgery. However, completely isolated duplication cysts have their own blood supply because of which the surgical resection of the cyst does not compromise the bowel vasculature.^[9, 10] Menon et al. suggested that a cyst with no discernible communication or connection to the adjacent alimentary tract but the presence of typical histopathologic features of a duplication cyst would qualify for the diagnosis of an isolated duplication cyst.^[11] The enteric cyst in our case was an isolated one with no connection to the main ileal lumen. Complete cyst resection was adequate in our case as there was no shared blood supply.

The imaging modalities commonly used for the diagnosis of duplication cysts are ultrasonography and barium studies. Double wall sign or muscular rim sign (Gut signature sign) seen on ultrasound imaging is characteristic though not specific of enteric duplication cysts. “Double wall” is characterized by

an echogenic inner rim which represents the mucosa surrounded by a hypo echoic rim corresponding to the smooth muscle layer. The double wall sign can also be seen in cases of mesenteric cyst and cystic teratoma. This sign can be completely lost in cysts with superadded infections.^[12]

CT, MRI and Tc99 are also useful modalities, with Tc 99 pertechnate scan being particularly useful for the detection of ectopic gastric mucosa with a sensitivity of up to 75%.^[6] MRI findings can support the sonographic findings, but may not give additional information on characterizing the cysts.^[13]

Histopathological examination confirms the clinical diagnosis of an enteric duplication cyst. The three essential histological microscopic features are: (1) a well-developed smooth muscle coat; (2) mucosal lining found within some portion of the alimentary tract; (3) contiguity to any segment of the alimentary tract.^[5] The mucosa is often similar to that of the region of the bowel to which the cyst is attached but at times some heterotopic mucosa like gastric or pancreatic tissue can also be found which needs to be documented.

Symptomatic intestinal duplications may require urgent surgical intervention as secretion from ectopic gastric mucosa may lead to ulceration of the intestinal mucosa, massive bleeding or bowel perforation. Though no consensus has been reached on treatment of asymptomatic duplication cysts, surgical excision is highly recommended to avoid late complications like malignant change. Exploratory laparotomy may be indicated if the preoperative diagnosis is unclear.^[4] Small cystic or short tubular duplications are treated by segmental resection along with adjacent intestine. In the present study our patient underwent surgical resection as he was symptomatic. The postoperative recovery and follow up period were uneventful.^[3]

Excision should be considered in all cases wherever possible. The surgical approach varies depending on the location and type of cysts, with resection and

anastomosis being needed in some cases as per the operating surgeon's preference.^[2]

CONCLUSION:

The case presented highlights the importance of a rare congenital anomaly which can not only remain asymptomatic for long, but also present with nonspecific symptoms. For a completely isolated enteric duplication cyst surgical resection followed by histopathological examination for confirmation is adequate without the necessity of an extended bowel resection.

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